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The Adult With Congenital Heart Disease: Cardiac Catheterization as A Therapeutic Intervention

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The revolution wrought by interventional cardiac catheterization in the management of congenital heart disease is reflected by both volume statistics and the number of currently "accepted" procedures (1). From a total of 10 cases in 1983, we now perform >700 interventional catheterization procedures (including a minority of adults) among 1,400 total catheterizations at the Children's Hospital, Boston. In addition, >12 new procedures have been added to the list of generally accepted procedures at our hospital.

Of the procedures that have become well established at our institution, five are of particular relevance to the adult with congenital heart disease: balloon dilation of congenital aortic valve stenosis, umbrella closure of atrial septal defect, occluder implantation for patent ductus arteriosus, umbrella closure of ventricular septal defect and umbrella closure of a silent patent foramen ovale for stroke prevention.

Dilation of congenital aortic stenosis. We have dilated >200 congenitally stenotic aortic valves in children beyond the neonatal period. The procedure has proved effective and extraordinarily safe. Gradient reductions of >50% have been achieved in 88% of the patients. Severe aortic regurgitation requiring surgical management has been a complication in <2%. There have been no deaths or strokes and the major morbidity (namely, vessel damage) has been reduced by the advent of newer catheters. Of considerable importance, restenosis has been extremely rare (outside the neonatal period) and <5% of patients have required redilation or surgery in the past 6 years.

These results contrast sharply with experience in adults with calcific aortic valves, in whom restenosis is very common, occurring in the majority of patients within 2 years. Stenotic valves are presumably degenerative trileaflet rather than congenitally abnormal. With respect to adults with bicommissural (bicuspid) congenitally abnormal valves, we have just reviewed the data and concluded that young adults with mobile noncalcified bicommissural valves will have the same overall results as will children. It seems likely that balloon dilation will assume increasing importance in the management of the young adult with congenital aortic valve stenosis.

Occluder implantation for patent ductus arteriosus. Catheter closure of a patent ductus was described by Portsmann et al. in 1971 (1). Current closure rates in properly selected patients range from 72% to 100%, with a low incidence of complications. Relevant to this discussion are two categories of adults with patent ductus arteriosus: 1) the young adult who is at risk of infective endocarditis because of a restrictive conical ductus focally narrowed at its pulmonary artery insertion, and 2) the older adult at hemodynamic risk because of a calcified, moderately restrictive ductus with the same morphologic features. In the younger patients, catheter occlusion avoids thoracotomy and eliminates the only risk confronting the patient—infective endocarditis. In the older patient, a hemodynamic burden is eliminated without the risk of surgery for a calcified ductus.

Umbrella closure of atrial septal defect. Although this procedure is 20 years old, the first widely successful device is the newly described clamshell umbrella. Nearly 100 atrial septal defects have been closed in adolescents or adults. In general, patients are selected on the basis of transesophageal echocardiographic findings. Atrial septal defects <20 mm in diameter are chosen if they are central and have an adequate rim of tissue (about 60% of patients are good candidates for umbrella closure). The procedure is performed with fluoroscopic or echocardiographic guidance: once the umbrella is correctly seated, it is released. Endothelialization (based on animal studies) is believed to occur within a few weeks, and the patients are managed for 6 months with aspirin alone. Complications have been few. Embolization (occurring early in the learning curve of a center) has been rare (2% to 3%) and asymptomatic but has required surgical removal of the device in three cases. Complete closure (absent shunt) occurred in 96% of cases with no late deaths, strokes or arrhythmias. The benign nature of this procedure for appropriate patients and the high success rate despite its novelty predict that it is likely to become the treatment of choice within a few years.

Ventricular septal defect closure. The most common indication for cardiac catheterization of the adult with congenital heart disease is the need to assess residual defects or dysfunction after "repair" of complex congenital heart disease rather than the diagnosis of simple congenital defects. A typical example is a patient who has a residual ventricular septal defect after reparative surgery. Such patients frequently have undergone several attempts at closure, have ventricular dysfunction and arrhythmias and are generally poor candidates for reoperation. We have developed a technique for transcatheter closure that has been successfully used to close nine ventricular septal defects in

adults with previously repaired congenital heart disease. The technique is among the most difficult of the catheter procedures, but it has been clinically successful in eight of nine defects with little morbidity.

Patent foramen ovale closure for stroke prevention. The advent of transesophageal contrast echocardiography has confirmed that a patent foramen ovale can set the stage for embolic strokes of "unknown" origin. Although we have closed only 12 patent foramen ovales for stroke prevention, none of the patients has had a subsequent stroke. The simplicity of the procedure raises the likelihood that heart "defects" previously thought to be benign will be managed by transcatheter techniques. The procedure is especially considered in young adults who present with stroke of unknown cause and who have a documented right to left shunt (by contrast echocardiography) through a patent foramen ovale.

Summary. The adult with congenital heart disease who undergoes cardiac catheterization at the present time is most likely to have complex heart disease and is left with clinically important sequelae or residual defects, ventricular dysfunction or arrhythmias. Residual defects such as paravalvular leaks, coronary fistulas and pulmonary artery narrowings may be corrected with transcatheter techniques. Patients with simple forms of congenital heart disease (for example, atrial septal defect, patent ductus arteriosus, aortic valve stenosis, pulmonary valve stenosis) will go to the catheterization laboratory for treatment, not diagnosis. Certain lesions previously considered benign (for example, patent foramen ovale) may require definitive interventional therapy to reduce the risk of stroke from paradoxic embolism.

Reference

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Electrophysiologic Residua and Sequelae

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Incidence and treatment of postoperative arrhythmias. With few or no exceptions, patients who undergo intracardiac surgery risk the development of postoperative arrhythmias and conduction defects (1). The highest incidence of arrhythmias occurs after intraatrial repair of complete transposition of the great arteries, intracardiac repair of tetralogy of Fallot and the Fontan procedure (Table 1). Electrophysiologic abnormalities may precede cardiac surgery, with arrhythmias reflecting the natural history of the congenital lesion.

Postoperative ventricular tachycardia is more likely to occur in response to the effects of long-standing right ventricular hypertension, healing of ventriculotomy scars under pressure, postoperative volume overload of the right ventricle and residual left ventricular pressure overload (Table 2). Ventricular arrhythmias are believed to be responsible for sudden death in 5% to 10% of patients with susceptible postoperative substrates.

Supraventricular arrhythmias are more common in lesions requiring extensive intraatrial surgery (complete transposition of the great arteries with intraatrial repair), especially when there is coexisting postoperative elevation of right atrial pressure as in the Fontan repair. These arrhythmias are associated with a 2% to 8% incidence of sudden death.

In patients whose cardiac lesion is associated with a significant incidence of arrhythmias, conduction defects or sudden death, meticulous postoperative surveillance is required, including electrocardiography, 24-h ambulatory monitoring and exercise stress testing when age permits. Electrophysiologic studies should be performed in patients with documented life-threatening arrhythmias and in high risk patients in whom such studies hope to uncover latent life-threatening arrhythmias, setting the stage for antiarrhythmic therapy. Postoperative inducible sustained ventricular tachycardia in patients with tetralogy of Fallot does not necessarily correlate with sudden death. Antiarrhythmic therapy is indicated in postoperative patients when a tachyarrhythmia compromises cardiac function or has a significant

Table 1. Incidence of Postoperative Arrhythmias

Congenital Heart Defect	Incidence of Arrhythmias (%)
d-transposition of the great arteries (intraatrial repair)	>50
Tetralogy of Fallot	30
Fontan repair	25
Ventricular septal defect	10
Atrial septal defect (secundum)	9